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An Incidental Finding of Libman-Sacks Endocarditis in a Young Female With Systemic Lupus Erythematosus Who Presented With Pleuritic Chest Pain

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Abstract

Libman-Sacks endocarditis (LSE) is a rare disease found incidentally in postmortem autopsies, characterized by microscopic to large verrucous vegetation on the cardiac valves, the most affected site is the mitral valve followed by the aortic valve. Females of reproductive age were observed as the most affected individuals as found in studies. Most individuals with LSE are asymptomatic and generally discovered lately when they presented with thromboembolic disorders such as stroke, cognitive disabilities, and death. Malignancy and autoimmune diseases involving systemic lupus erythematosus (SLE) and antiphospholipid syndrome (APS) are considered the primary etiology of LSE. As recognized, the majority of LSE cases are asymptomatic, it tends to be challenging to spot the condition at the early pathway of the disease. In this paper, we describe a young female who is known to have SLE on medications, she presented to the emergency department (ED) due to chest pain and exertional dyspnea for a few days, laboratory investigations showed anemia, raised inflammatory marker, and anti-DsDNA. Imaging studies showed bilateral pleural effusion on the chest X-ray and a large vegetation on the posterior mitral valve with moderate regurgitation and normal wall motion in transesophageal echocardiography. The patient was managed by pulse steroid therapy, anticoagulation therapy, and a low dose of diuretic, the patient improved dramatically and discharged home with close follow-up in the clinic. The primary treatment of LSE is anticoagulant therapy, however, surgical intervention should be considered in case of large vegetation recurrent thromboembolism despite anticoagulant therapy. As the prognosis in LSE is considered very poor and there is no definitive laboratory investigation exists to confirm the diagnosis, we highlight the importance of considering LSE as a serious and crucial differential diagnosis when dealing with SLE patients who presented with dyspnea and pleural effusion secondary to valvular dysfunction, mainly the mitral valve.

Categories: Internal Medicine, Rheumatology, Medical Education

Keywords: mitral valve vegetation, anticoagulant therapy, warfarin, systemic lupus erythematosus, libman sack endocarditis

Introduction

Systemic lupus erythematosus (SLE), one of the most prevalent chronic relapsing inflammatory autoimmune diseases, affects several organ systems and manifests clinically in a variety of ways [1]. According to estimations, there are 3.41 million patients with SLE worldwide, with a prevalence of 43.7 (15.87 to 108.92) per 100,000 people. United Arab Emirates had the highest estimated incidence of SLE (166.92, 139.01 to 198.54 per 100 000 people) in the world [2]. Most studies found that the incidence of SLE in females peaks in the middle and younger age, whereas the peak incidence for men often occurs later, between the fifth and seventh decades [3]. Despite the enhanced chance of survival, SLE patients' mortality rates are estimated to be 1.4-5 times higher than those of the general population [4]. As was observed in many studies, the survival rates at 5, 10, and 15 years after the diagnosis were 96%, 93%, and 76%, respectively [5].

SLE individuals may have a variety of systemic symptoms, ranging from general symptoms, such as fever, malaise, myalgia, and weight loss, to specific systemic manifestations involving cutaneous, musculoskeletal, renal, and cardiovascular organ systems [6]. Cardiovascular involvement in SLE results from a synergistic combination of pathogenic processes, resulting in the development of many cardiac events at a younger age than the general population. The most prevalent cardiac outcomes in SLE patients are pericarditis, myocarditis, valve disorders, and conducting system disorders [7].

According to estimations, one in every 10 SLE patients had Libman-Sacks endocarditis (LSE). LSE is a sterile lesion that occurs concurrently with SLE and antiphospholipid syndrome (APS) targeting the mitral and aortic valve, causing stenosis or regurgitation [7]. A hypercoagulable condition is commonly linked with nonbacterial thrombotic endocarditis such as in solid tumors, SLE, and APS [8]. LSE patients are usually asymptomatic, however, they might have a variety of presenting symptoms, in terms of arrhythmia, pericarditis, and heart failure secondary to mitral valve regurgitation, but according to data, valve

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vegetation increases the risk of embolic cerebrovascular disease, which they might present with ischemic stroke or transient ischemic attack (TIA) [8,9].

We reported a young female who had SLE presented to ED because of chest pain and shortness of breath and was incidentally found to have a large thrombus versus vegetation on the posterior mitral valve through transesophageal echocardiography. Finally, the patient was labeled to have LES and started on anticoagulant therapy. The aim of the report is to expand the knowledge, awareness, and importance of early detection of LSE in SLE patients to prevent cerebrovascular and cardiovascular complications and disabilities.

Case Presentation

We report a 17-year-old Saudi female, a single, student, known to have systemic lupus erythematosus disease (SLE). In 2020, her initial presentation was a butterfly rash on her face, arthralgia in the small joints of her hands, and hair loss. The antinuclear antibody (ANA) and anti-DsDNA were positive. The SLE diagnosis was confirmed and prednisolone and hydroxychloroquine were initiated as a therapy.

Two years apart from the disease diagnosis and remission state, in late August of 2022, the patient started to have gradual left-side chest pain for four days with strict compliance to her home medications, the pain is not radiating or referring to any site of the body, associated with exertional dyspnea and dry cough. After four days, the patient came to the emergency department with that complaint and was examined. Up on examination, the patient was conscious, alert, oriented, and not in respiratory distress, but in mild-moderate pain. Vitals, she was afebrile at 37.4°C, heart rate 110 bpm, respiratory rate 20/minute, blood pressure 135/65 mmHg, and oxygen saturation (96% on room air). On oral cavity exam, good hygiene, and no ulcer. During the neck exam, no swelling or lymphadenopathy was appreciated. Chest auscultation reduced air entry bilaterally in the base of the lungs and other chest exams were unremarkable. On cardiovascular examination, jugular venous pressure (JVP) was elevated at about 11 cm. On auscultation, the patient had normal first and second heart sounds with no added sounds, and no parasternal heave was appreciated. Abdominal examination revealed a soft, non-tender abdomen, and no hepatosplenomegaly. In lower limb examination, patent peripheral pulses, grade +1 edema in both legs, no erythema, and negative signs for deep vein thrombosis.

Laboratory and imaging studies

On arrival at the emergency department (ED), blood samples were taken, which showed low hemoglobin 7.1 g/dL, total white cells $5.5 \times 10^9/L$, lymphocyte count $0.36 \times 10^9/L$, high anti-DsDNA antibody, high ESR 137 mm/hr, CRP was 11.8 mg/dL, and normal urea/creatinine level. C3 and C4 were low. Septic work was sent for three sets of blood cultures, and it became negative for organisms. The electrocardiograph showed a normal sinus rhythm with low voltage on limb leads.

The chest X-ray showed bilateral pleural effusion with a flask cardiac silhouette (Figure 1). A computerized tomography (CT) scan conducted and was showed bilateral pleural effusion with pericardial effusion (Figure 2).

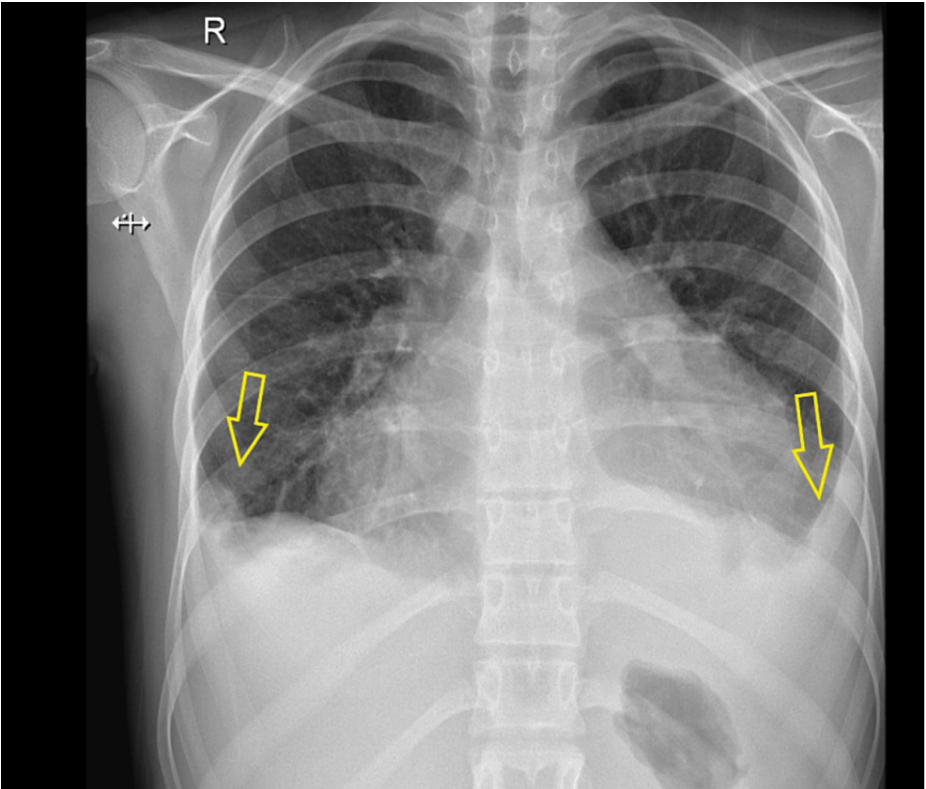


FIGURE 1: Initial chest imaging before initiating the management plan

Image description: posterior-anterior view of the erect chest X-ray showing cardiomegaly with a silhouette sign and bilateral obliteration of costophrenic angle more in the left side, which goes with bilateral pleural effusion.

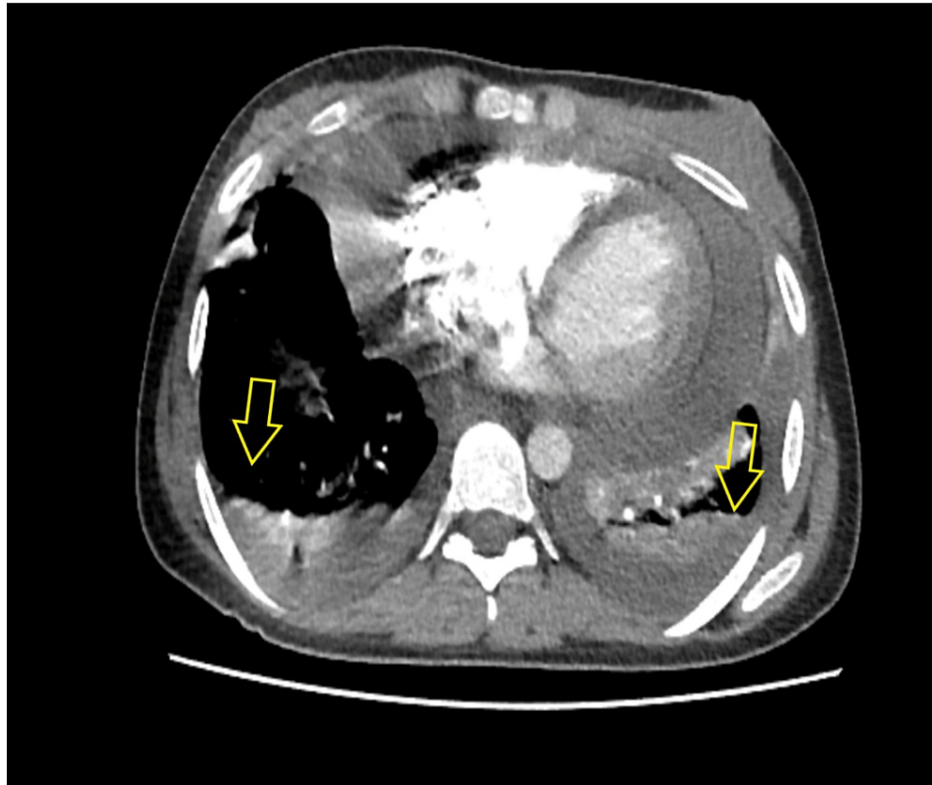


FIGURE 2: A cross-sectional computer scan study with contrast showing pericardial effusion with passive lung atelectasis more on the left side

Lung parenchyma shows a diffuse bilateral mosaic attenuation pattern.

Transesophageal echocardiography (TEE) was requested urgently, and it showed a bulky mass/thrombus on the posterior mitral valve about 8 x 18 mm, the left ventricle was normal in size and systolic function, the right ventricle was normal in size and function as well (Figures 3-4). The TEE findings were highly suggestive of LSE. An abdominal ultrasound was requested and showed minimal ascitic fluid.

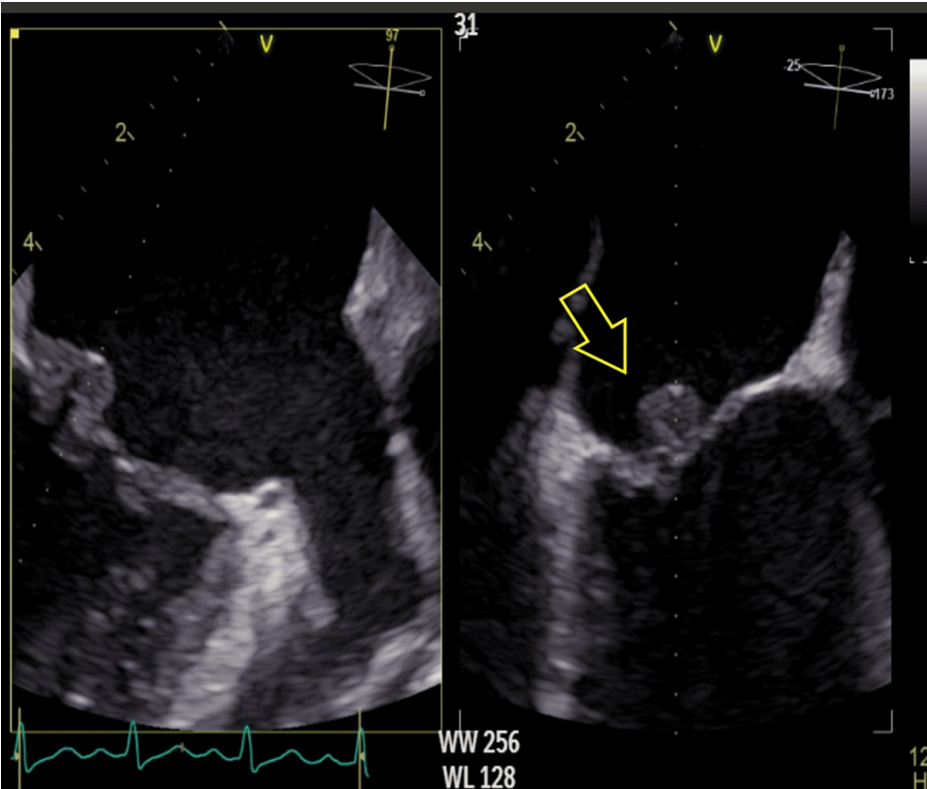


FIGURE 3: Transesophageal echocardiography shows a bulky thrombus/mass on the posterior mitral valve leaflet measuring 8 x 18 mm

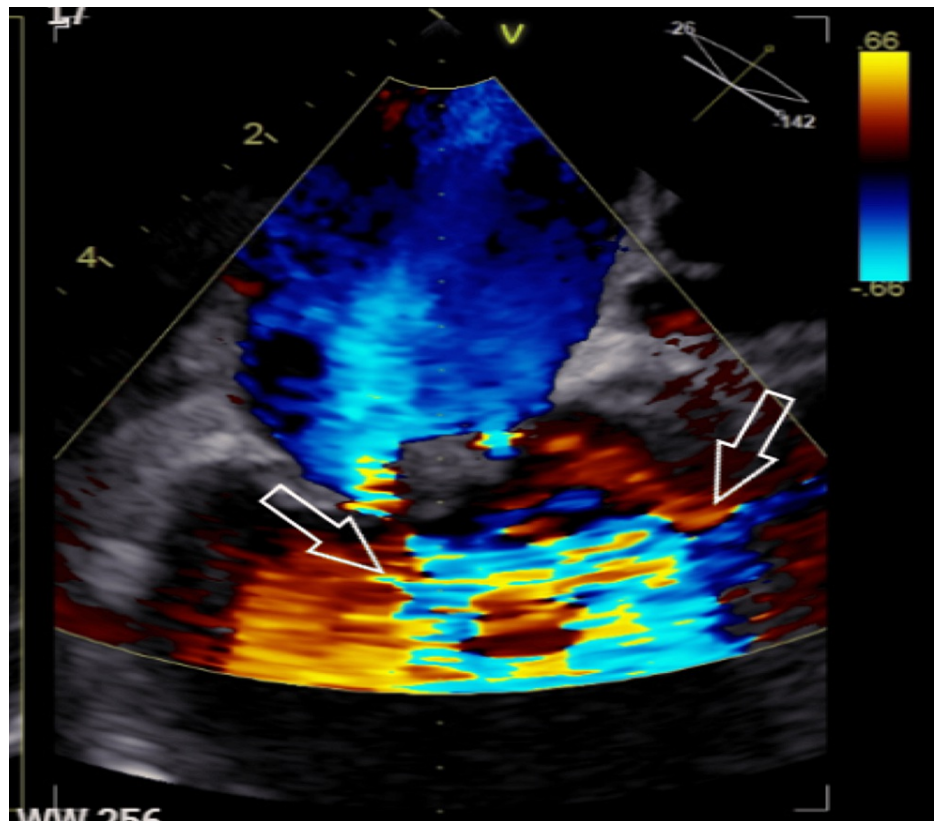


FIGURE 4: Transesophageal echocardiography shows moderate mitral regurgitation

The patient was admitted to the regular ward with an SLE flare in terms of serositis and LSE for pulse steroid therapy and therapeutic anticoagulation. The patient was started on methylprednisolone as a pulse therapy, bridging the anticoagulation information of low molecular weight heparin to warfarin and a low dose of furosemide 40 milligrams intravenous once daily for three days. Five days later, the patient showed a significant improvement after receiving pulse steroid therapy and therapeutic anticoagulation with no complications (Figure 5). The patient was discharged home safely on a tapering dose of prednisolone (2 mg/kg) and warfarin with close outpatient follow-up within three days for INR monitoring and within two weeks for SLE flare.

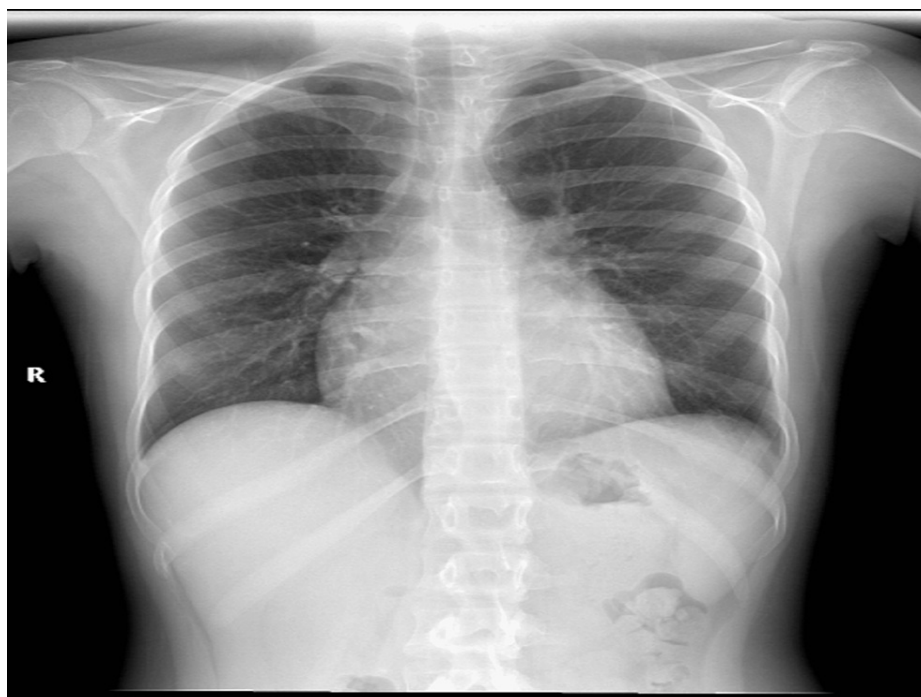


FIGURE 5: Chest X-ray repeated after five days of pulse steroid therapy and a low dose of diuretics, which shows a huge improvement in comparison to the previous one, in terms of improvement in the pleural effusion and cardiomegaly

Discussion

A variety of cardiac and vascular complications can develop in SLE patients, affecting the pericardium, myocardium, valves, coronary arteries, and conduction system. LSE is an uncommon disorder with prevalence in an autopsy series, ranging from 0.9% to 1.6% that is most frequently discovered post-mortem [10,11]. In the demonstrated case, the primary etiology for the verrucous vegetation was SLE; interestingly, many studies of autopsic cases who are found to have LSE are more often diagnosed with concomitant cancer, which reflects that SLE is not only the primary etiology of LSE, cancer as well can cause verrucous vegetation in non-SLE patients. On the other hand, patients with a history of previous cardiac and valvular surgery were more likely to have IE, which increases the risk of bacterial or fungal vegetation [12].

The majority of SLE patients who have LSE are asymptomatic; however, LSE individuals are more likely to present with complications of LSE such as cerebrovascular embolism (stroke or transient ischemic attack) and systemic embolism (mesenteric, limb ischemia, or peripheral arterial embolism) [8]. As described in the previous studies, many individuals who have LSE are either asymptomatic or present with thromboembolism manifestations. However, our patient presented with heart failure manifestations with preserved ejection fraction secondary to SLE flare in terms of serositis and moderate mitral valve regurgitation with verrucous vegetation. There are few studies in Saudi Arabia that were documented and published for SLE patients who presented with heart failure symptoms secondary to an SLE flare in terms of serositis and LSE with preserved ejection fraction.

The diagnosis of LSE in SLE patients is challenging, yet there is no definitive laboratory test to confirm the diagnosis of LSE. A high clinical suspicion of LSE will lead to the diagnosis, especially in SLE patients who have pleural effusion either secondary to SLE flare in terms of serositis or mitral valve regurgitation secondary to LSE, a full assessment should be done with laboratory studies. The most important two investigations in LSE are three blood cultures before starting empirical antibiotics and transoesophageal echocardiography. A verrucose vegetation in LSE has specific characteristics, mainly irregular borders, absence of independent motions, heterogeneous echo density, and involving the basal and mid-portion of the mitral and aortic valves [8].

The core management of LSE is systemic anticoagulation regardless of the underlying cause (malignancy or SLE). Low molecular weight heparin (LMWH) or unfractionated heparin at therapeutic doses should be utilized instead of factor Xa inhibitors (such as apixaban, rivaroxaban, and dabigatran) if APS has not been ruled out. On the other hand, in the setting of autoimmune or inflammatory disorders, warfarin can be used with an extended survival rate in LSE [13]. All individuals who receive anticoagulant therapy, especially on

warfarin must be educated in a written order for drug interaction and the need for regular follow-up in an anticoagulant clinic. In our presenting case, the patient presented with SLE flare-up in terms of serositis with pleural effusion, chest pain, and tachycardia with low complement serology, started on pulse methylprednisolone therapy for three days, therapeutic LMWH and furosemide for pleural effusion, the patient showed a huge improvement and was discharged home safely on mycophenolate, hydroxychloroquine, prednisolone, and warfarin with close follow-up. Pulse steroid therapy should be considered in severe SLE flares (e.g., central nervous system, renal system) as induction therapy for rapidly controlling the flares and reducing inflammatory process, however, minimizing exposure must always be made to prevent long-term complications (opportunistic infection, gastric ulcer, and osteoporosis) [14].

As reviewed in many studies, valvular vegetation and regurgitation are seen in up to 61% of SLE patients. Surgical intervention in LSE has a crucial role and a strong indication in case of giant vegetation and recurrent embolization despite therapeutic anticoagulation. A mechanical valve is preferred among young patients with APS who require lifelong anticoagulants. The risks and benefits should be weighed while considering surgery in relation to the underlying cause and life expectancy [15].

Conclusions

LSE is a fatal clinical condition if it is not discovered early. As most of the individuals are asymptomatic, they mainly present with complications of LSE such as stroke, mesenteric ischemia, or limb ischemia. Since there are no laboratory tools to make a definitive diagnosis of LSE, high clinical judgment and suspicion need to be considered in individuals with SLE who presented with either pleural effusion secondary to mitral valve regurgitation and vegetation or in late complications such as thromboembolic events. Transesophageal echocardiography has a crucial role in diagnosing LSE. The core treatment of LSE is anticoagulant therapy. However, surgical intervention can be considered in case of recurrent thromboembolic events and in the presence of large vegetation. The prognosis is poor, as LSE individuals are prone to recurrent thromboembolic events, cognitive disability, and death. The aim of this case report is to expand the knowledge and awareness of early detection and management of LSE in terms of preventing further complications and disabilities.

Additional Information

Disclosures

Human subjects: Consent was obtained or waived by all participants in this study. **Conflicts of interest:** In compliance with the ICMJE uniform disclosure form, all authors declare the following: **Payment/services info:** All authors have declared that no financial support was received from any organization for the submitted work. **Financial relationships:** All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. **Other relationships:** All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

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